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CORRESPONDENCE

Sclerodermiform lupus erythematosus: A rare case presented as cicatricial alopecia

Dear Editor,

Chronic cutaneous lupus erythematosus (CCLE) is associated with interface dermatitis with hydropic degeneration of the basal cell layer. Along with lichen planopilaris, it is the most common cause of inflammatory primary cicatricial alopecia.[1] Localized scleroderma, or morphea, is characterized by an excessive collagen deposition leading to thickening of the dermis and/or subcutaneous tissues. The coexistence of CCLE and localized scleroderma in the same lesion is exceedingly rare. The term *sclerodermiform lupus erythematosus* thus was proposed.[2–5] We herein describe a patient with features characteristic of sclerodermiform lupus erythematosus, which presented as a cicatricial alopecic plaque on the scalp.

A 33-year-old woman with no relevant medical history presented herself with chief complaint of hair loss. Upon inspection, we found a palm sized, slightly depressed indurative plaque associated with near-complete hair loss and telengiectasia on her vertex scalp. Follicular plugging could be seen upon close examination.

Two 4-mm punch biopsies were performed. Histologically, diffuse whole-layered dermal sclerosis involving dermosubcutaneous junction, extended into the subcutaneous septa and destructed hair follicles. Superficial and deep, periadnexal and perivascular lymphoplasmacytic infiltrates were observed. Perifollicular inflammation, thickening of basement membrane and focal concentric lamellar fibroplasia were seen. The epidermis revealed atrophic change with focal basilar vacuolar change. Alcian blue stain revealed focal dermal mucin deposition. The picture was compatible with lymphocytic cicatricial alopecia, which showed overlapping feature of both CCLE and morphea.[2–4]

Concurrence of clinical and histopathological features of both CCLE and localized scleroderma in the same lesion was first described in 1978,[5] and termed *sclerodermiform lupus erythematosus*.[2,6] The finding is exceedingly rare, and almost all the cases described were female. Most lesions are located on the limbs and are distributed along the Blaschko's line.[2,3] One proposed etiology is an antigenic skin mosaicism manifestation triggered by exogenous stimulus, such as viral infection. An inflammatory T-cell response caused by the exogenous trigger may results in tissue damage limited to the areas carrying the genetic mosaicism.[3] Another possible explanation is immunocompetent maternal cells passed through the placenta during pregnancy, inducing a chronic graft-versus-host disease-like reaction solely in areas with skin mosaicism.[3,7] As for the etiology of the *overlap* condition, some proposed that the initial lichenoid inflammation or keratinocyte apoptosis of the CCLE component acts as the *trigger* event, which in turn provokes abnormalities in fibroblast-dependent reparative mechanisms and eventually leads to tissue sclerosis.[2,3]

The most commonly prescribed therapies are systemic antimalarials, combined with topical or intralesional steroid.[2,3] Recognition of this cutaneous overlap syndrome help us to provide better treatment advice, based on the well-recognized beneficial effect of antimalarials at least on the CCLE component (Figures 1 and 2).[8]

Figure 1 A 7 cm × 2.5 cm alopecic plaque on the vertex with loss of follicular ostia.

Conflict of interest: The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in this article.

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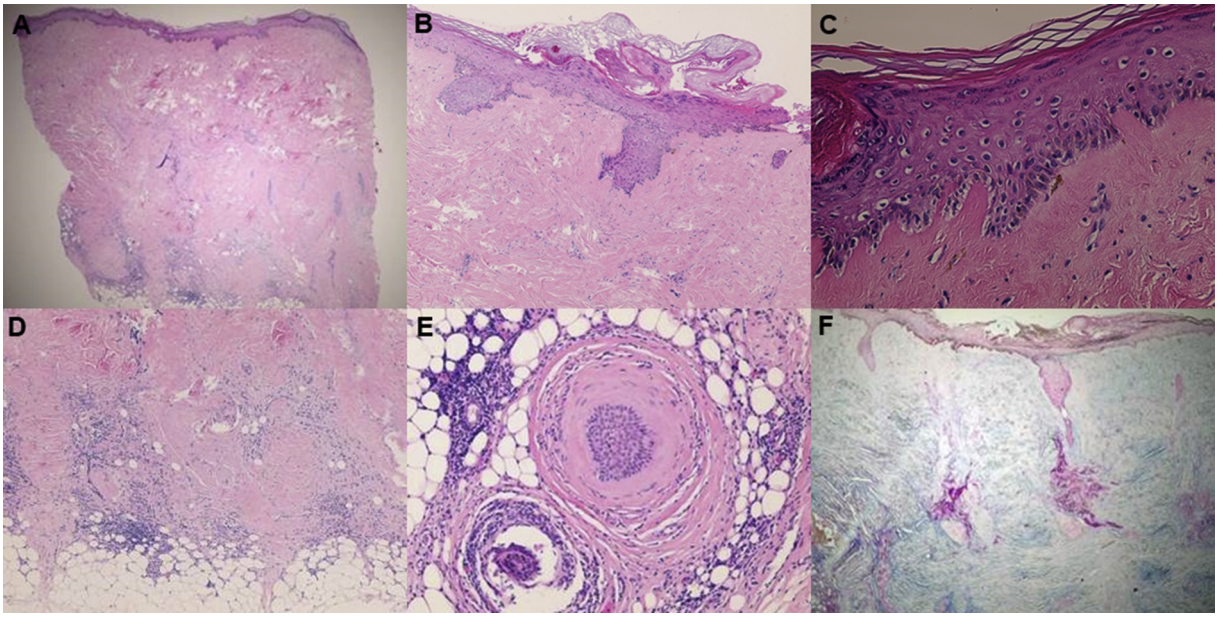

Figure 2 (A) Follicular plugging of the epidermis, periadnexal inflammation, near-complete loss of follicular structures and diffuse dermal fibrosis [hematoxylin–eosin H&E) stain, original magnification: × 40. (B, C) Higher magnification shows interface dermatitis with vacuolar change at the dermal–epidermal junction (H&E: B ×100, C ×200). (D) Higher magnification shows lymphocytic infiltration and destruction of hair follicles. Significant fibrosis involved the dermosubcutaneous junction and also extended into the subcutaneous septa (H&E, ×100). (E) Horizontal section shows dense perifollicular lymphoplasmacytic infiltration, concentric fibrosis and destruction of hair follicles (H&E, ×200). (F) Focal presence of dermal mucin (Alcian blue, ×80).

Yu Yu*
Department of Dermatology, Cathay General Hospital, Taipei, Taiwan

Chih-Yi Liu
Department of Pathology, Cathay General Hospital, Taipei, Taiwan

Jung-Yi Chan, Feng-ling Lin
Department of Dermatology, Cathay General Hospital, Taipei, Taiwan

* Corresponding author. Department of Dermatology, Cathay General Hospital, 280 Renai Road, Section 4, Taipei, Taiwan.
E-mail address: a0982304275@gmail.com (Y. Yu).

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